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ABSTRACT

This paper comprises a compilation of nine case studies of girls (aged 4-16 years) with Rett Syndrome. The educational settings involved are various and include private day school, public elementary school in both integrated and special needs classrooms, and a county-operated preschool program for handicapped children. Each case study outlines the student's behavior, including such behaviors as the hand wringing common with Rett Syndrome children, hyperventilation, breath-holding, the ability of some to walk independently, etc. Suggested educational and therapeutic intervention activities that have been found effective are described, such as total communication approaches, hydrotherapy, bubbles, musical instruments, physiotherapy, eye-pointing, and cause and effect activities. (JDD)

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SUGGESTIONS FOR EDUCATIONAL AND THERAPEUTIC INTERVENTIONS WITH THE RETT SYNDROME CHILD

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Every child with Rett syndrome has educational and therapeutic needs that are unique to her situation. The strategies described in this text are those of trained individuals who have had experience with the Rett syndrome child.

The International Rett Syndrome Association does not recommend a particular treatment or therapy.

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Aimee

Submitted by Jamie Murphy, B.S.
Severe Special Needs Teacher
The I.S.E.C. School
The University Hospital
Boston, Massachusetts

Aimee is an 8 year-8 month old girl who has attended The I.S.E.C. School since November, 1983. Aimee has been diagnosed by her neurologist as "a youngster with probable Rett syndrome". Developmental regression was noted at 20 months.

I.S.E.C. is a private day school which Aimee attends 5 days a week, 12 months a year. Aimee is in a class with 5 other students, one of whom has also been diagnosed with Rett syndrome. There is a 1:2 staff/student ratio in Aimee's classroom.

Aimee walks independently, but hesitates at points where the elevation or surface changes and may need physical assistance in these situations. Aimee also needs time to adjust to changes in position and movement (for example: if sitting, Aimee needs time to make a transition to a standing position). Aimee has a great deal of difficulty walking after sitting for long periods of time (i.e. bus ride to school). Thus, Aimee's school day is scheduled to provide her with alternating periods of sit-down activities and movement activities. In addition, Aimee cannot always remove herself from dangerous situations (i.e. another child pulling her hair). Aimee receives daily half-hour physical therapy sessions and range of motion exercises.

Aimee uses her left hand, however, due to Aimee's "hand wringing" behavior, she occasionally needs physical assistance to separate her hands. Aimee is often able to separate her hands spontaneously, especially when motivated (i.e. water play). Aimee independently eats finger foods, given support of her arms by a tray placed on the table at chest level. Aimee requires physical guidance or complete physical assistance for spoon feeding, cup drinking, dressing and undressing. Aimee uses a small cut-out tumbler and a bent handle spoon with a cuff at this time. Aimee is being scheduled to be trained at this time. She is on a 1 to 2 hour toileting schedule and wears training pants at school. She has a 65% success rate (dry at toileting) at this time.

Aimee learns best in individual and small group sessions with a familiar instructor. She requires a total communication approach including the use of pictures, gestures and verbal requests. Communication is an emerging skill for Aimee. She is able to express her needs and choices through the use of pictures, direct selection, vocalizations and facial expressions. Aimee is able to make choices by touching pictures, reaching for or walking toward preferred objects, people or activities. She responds to simple gestural/verbal commands such as "come here" and "stand up". She also

responds to her name. Aimee indicates awareness of familiar people (especially her mother) by smiling and walking toward the person.

Aimee engages in hand-wringing behavior which interferes with functional hand use. She has periods of breath holding and blank staring which limit her ability to attend or respond. Also - Aimee is often lethargic and hard to motivate in the morning. Aimee takes Tegretol for seizure control.

Aimee enjoys interacting with adults and other students. She responds well to enthusiastic verbal praise. Aimee's play skills have been improving recently. She will independently and enthusiastically engage in water play and ball throwing. Her play skills include operating Teddy Ruxpin with a press panel, batting at suspended balloons and pulling on a ring to move a car. Aimee also enjoys walks, cuddling and music.

Jennifer

Submitted by Jamie Murphy, B.S.
Severe Special Needs Teacher
The I.S.E.C. School
The University Hospital
Boston, Massachusetts

Jennifer ia an 8 year-3 month old girl who has attended The I.S.E.C. School since January 1985. She has been diagnosed as having Rett syndrome. Developmental regression was noted at 15 months.

I.S.E.C. is a private day school which Jen attends 5 days a week, 12 months a year. Jen is in a class with 5 other students, one of whom has also been diagnosed with Rett syndrome. There is a 1:2 staff/student ratio in the classroom.

Jennifer walks independently, but requires assistance on stairs. Jennifer often waits for an adult to take her hand, and this has increased recently. Jen does not enjoy sitting for long periods of time and thus has a schedule with alternating periods of sit-down and movement activities. Jennifer has daily 1/2 hour sessions of physical therapy and range of motion exercises at least 3 days a week. Jennifer recently began wearing a scoliosis jacket at night.

Jennifer uses her left hand functionally to eat finger foods, uncover hidden food, and touch favorite toys; however, she often needs physical assistance to separate her hands. Jennifer's ability to use her hand improves significantly when her arms are supported by a tray at chest level. She uses her right hand to stabilize herself and to protect herself when she is off balance. She can bring a cuff spoon or fork to her mouth once the food is on the spoon or fork. She will visually attend to hand-over-hand scooping or stabbing. She requires close physical guidance to drink with a two handled cup (hands slipped through handles, around cup). Jen needs total physical assistance for dressing and other areas of self care. She is on a toilet-schedule program. She wears training pants at school and is toilet trained every 1-1/2 to 2 hours. Her success rate (dry at toileting) is 76%.

A total communication system is used throughout the day with Jennifer. It employs the use of pictures, verbal statements and gestures. Jen is able to use pictures to make choices between foods and leisure activities. We are currently assessing which selection method Jennifer should use when communicating. The systems we are investigating are: direct selection with her left hand, eye gaze, head pointer and the possibility of utilizing computers and various switches. Jennifer is also able to express her emotions and desires through a combination of facial expressions, vocalizations, looking at, reaching toward, and touching objects and people. She is able to follow a few simple directions such as "come here" and "stand up."

Jennifer engages in stereotypic hand-clasping, breath holding and blank staring throughout the day. Though these behaviors interfere with educational programming, they appear not to be within her control. It is necessary to separate her hands and wait for regular breathing before continuing a task. Jennifer takes seizure control medication.

Jennifer is a social child who actively seeks out and initiates contact with both children and adults. She tends to explore her environment using her eyes more than her hands. She will walk up to pictures, toys and people and incline her head toward them. She enjoys looking at pictures. The use of Active Stimulation devices (A.S.P.) has proved to be very successful with Jen. She enjoys Teddy Ruxpin and will use a switch to turn him on. Although Jen will separate her hands when motivated (attaining food), she rarely separates her hands to manipulate or touch objects and toys in her environment. Food reinforcers and social praise are good motivators for Jennifer. Swimming is an activity Jen enjoys, she is able to independently keep herself afloat momentarily. When using a floatation device she is able to keep herself upright and afloat for 2 to 5 minutes. Jennifer also enjoys walks and music.

Katie

Submitted by Nancy Ericksen
School Association for Special Education
DuPage County, Manning School
Westmont, Illinois

Katie is a beautiful 4 1/2 year old girl. Her eyes are bright and expressive, her smile charming and she gives hugs by snuggling and leaning into your neck.

She attends a Multiple Needs Preschool program for children ages 3-6 years old whose primary disability is in the area of mental retardation and/or severe impairments in communication. The program heterogeneously groups students in a classroom located in an elementary school. The curriculum follows a functional model emphasizing chronological age-appropriate skills taught in the context of community and school environments.

Pregnancy was full term with no complications. Katie rolled at 9 months, walked at 18 months. She began to speak syllables at 11 months, but stopped after 1 year. She is not toilet trained, but has some success on an hourly time schedule. At first glance, Katie appears tense and rigid, however, she predominately lacks muscle tone. Her gait is stiff-legged and wide based. She cannot move from standing to a sitting position nor back without assistance. Her hands and feet are often cool. She is very thin - 21 lbs. The hyperventilation and breath holding patterns interfere with eating. She often falls asleep 15-20 minutes into a meal.

Katie seems to go through a "mood cycle" 2-3 times per day at school. She begins her day relaxed and smiling when she gets off the bus (hands still clasping) and becomes very agitated 1 1/2 - 2 1/2 hours later (crying, screaming, increased tenseness, hand clasping and breathing patterns). This pattern repeats. We are currently trying to find activities which have an effect on this cycle (i.e., speed it up or slow it down). When Katie gets tired she rubs the side of her face and hair. We immediately try holding her and rocking her slowly, and she will sometimes fall asleep in a beanbag chair. When she has reached the very agitated stage, and the holding and rocking has not quieted her, we have found that wrapping Katie as you would swaddle a newborn helps to reduce the tenseness and irritability. Her breathing almost instantly becomes more relaxed and the crying and screaming usually stops within 5 minutes. We use a sheet folded in half and gently wrap her, securing one hand away from the other. Within 10-20 minutes Katie is either asleep or ready for her next activity. We have tried keeping a food diary looking for patterns and possible allergies, but found none. We are currently keeping a record of Katie's activities and reactions.

When working on functional hand skills, we hold down her left hand. We have found only a few activities which motivate

Katie to use her hands. Katie enjoys reaching for food, bubbles, hitting a tambourine and most recently, pressing a switch to turn on a tape recorder (as described by Jackie Levin, IRSA Newsletter, Winter 86). The bubbles and tambourine need to be held about 6-8 inches directly in front of her eyes. She can hit the switch when it is on a table in front of her. She has just begun to respond to music by smiling.

We are most excited about the following technique which has given Katie a way to communicate. At snack and lunch Kate requests a specific food item from a field of 2 choices (77% of the time). These are held one over the other (rather than next to each other), 8-12 inches in front of her eyes. She makes her choice by eye pointing, reaching or both. Katie's reach is a general arm swing or swat at the food, and when the food items are held side by side (horizontally), she hits the first item closest to her arm. When the items are held one over the other, she makes definite choices - sometimes the top item and sometimes the food on the bottom. Given a choice between a cookie and a spoon with jello, Katie will consistantly choose the cookie, whether it is in the top or bottom position.

The above activities were developed in a cooperative effort (transdisciplinary approach) of her teacher, ancillary staff, and of course, Katie's parents.

Tanya

Submitted by Sharon Bewza
Rockland Elementary School
Campbell River, B.C.
Canada

Tanya is a 7 year old girl who attends a Special Needs Class in a public elementary school. She is in this class with 6 other moderate-severely mentally handicapped children ages 5-9. Along with the Special Education teacher are three full-time aides, one of which is assigned to Tanya.

Tanya has most of the characteristics of Rett syndrome. Fortunately, she has not had any seizures, nor does she suffer from any eating or digestion problems (prior to Tanya's diagnosis last year, she was on a restricted diet because her doctor felt perhaps she had a severe allergy problem. Tanya now eats most foods, however, with no apparent problems.) Tanya does not feed herself. She is ambulatory, but is easily knocked off balance by other children (or adults!). She functions well below a 1 year old level. Sometimes Tanya is irritable and 'teary' but after a short nap she usually returns to her happy 'chatty' self. Tanya's regression began at approximately 17 months of age.

SUCCESSFUL THERAPIES

A. Eye Contact

- 2 year program: reward was food at first, and later music
- success rate 60%
- spontaneous eye contact has increased in frequency and length
- we now give lots of praise and/or hugs for eye contact.

B. Carrying her lunch kit to the table:

- began with hand over hand
- after placing the lunch kit in the left hand (she just dropped it if placed in her right hand). Tanya will now carry it quite independently. Occasionally she requires a touch on her shoulder to keep her walking.
- we are now encouraging her to carry other objects.

C. Physiotherapy

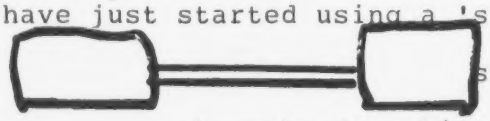
1. Vestibular swing: Tanya has progressed from hand over hand hanging on to independent hanging on - she can balance well with a fair amount of applied swinging movement.

2. Fisher Price Corn Popper Toy: We lengthened the push stick and made the handle larger and easier for Tanya to grasp - again we started out hand over hand and now Tanya will push the Popper independently for 5-10 minutes. She really watches the 'corn popping'!

3. Large Ball - we say, "Tanya, touch" and give the ball a good push - this has helped increase her protective reaction.

D. Hydrotherapy

Tanya, along with her classmates, goes to the local pool twice a week. Her parents also built a jacuzzi recently, which Tanya really enjoys. We have seen improvement in Tanya's ability to hold on to the side of the pool for increasing lengths of time while putting her through a series of leg exercises. She is also tolerating being on her back for longer periods - especially if her left hand is held beneath her. We have just started using a 'styrofoam stick':



total length 3'

Tanya holds on very well and she can be pulled or pushed through the water.

E. Feeding Program:

Tanya has learned to eye point for a choice of up to 3 foods plus a drink. She has been consistently eye-pointing for 1 1/2 years. We started this program in a small, bare enclosed room and Tanya has just recently progressed to the point where she can eat near her classmates, but at a separate table and with her back to them. (Otherwise she doesn't stay 'on task'). We are now working with her on 'hands on hands' feeding after she eye points to what she would like.

To improve her eye-pointing ability we are planning to use a tape of her Mom, Dad and brother with corresponding pictures and - hopefully - she will connect the picture and the voice. From there, we hope she will eye point to the picture of the person whose voice she hears. We are hoping this will be the beginning of eye pointing communication.

We have restrained her left hand during any work or feeding session and find she is more relaxed and attentive. Tanya constantly wrings her hands when one or both are not restrained.

Tanya has very devoted parents (and a big brother) who have spent many countless hours working with her.

Ro

Submitted by Mrs. Anne Dobbelaere
Kindergarten Teacher
Edward Smith School
Syracuse, New York

Ro is a beautiful, ambulatory seven year-old child. She attends Ed Smith School in an integrated kindergarten. There are six special needs children and twenty-five typical students.

When I first saw Ro at her preschool setting, it was clear that programming for her would be a challenge. I started going to garage sales and toy shops to look for motivating items I could use with Ro. Ro enjoys Miss Piggy, Kermit, Sesame Street characters, musical instruments, records and cause and effect toys. Ro's program is planned around using high interest items. For example:

1. A small Miss Piggy finger puppet and a Kermit finger puppet helps establish object permanence.
2. Figures of Bert, Ernie and Big Bird are put in a container of water to encourage Ro to grab for them.
3. A large Miss Piggy monster was placed on the bathroom door so Ro would walk toward it.
4. A large Miss Piggy puppet is placed far enough out of reach so Ro will try to get out of her chair. Gradually she could pull up her own weight and get out on her own.

To increase Ro's ability to hold things, we started out with lightweight rattles and gradually increased their weight. At the beginning of the year, Ro could hold the rattle for three seconds. Her hold increased up to thirty seconds. We started using musical instruments like bells, tambourines, cymbals, and rhythm sticks. A Fisher Price tape recorder, Cookie Monster piano and musical birds all encourage purposeful use of her hands and are a means of communication.

Ro will tap at a picture of an item that she wants when given a choice of two pictures. Pushing a small plastic shopping cart or carriage increase Ro's grasp and walking.

Markers and paints (in bright colors) provide Ro with instant results. She holds the marker in a clenched fist and gets wrist support from an adult; sometimes she can do this independently after the marker is placed in her hand.

Ro pushes,bats, taps, squeezes, and pulls toys such as Fisher Price Rally, Polly chimes, apple, cow, and walking dog, Ernie squeeze toy, colorful ball and others.

There are various adapted toys and switch plates and music that we use at play time. Our library has a lending section of adaptive toys and a toy can be kept for two weeks.

Ro is in the process of being potty trained. At this time, we are working on communication so that the hourly trips to the bathroom can be eliminated. If she is asked and tells us no we take her at her word. Ro is about 80% trained.

A computer program is in the beginning stages. A special disc was made for Ro for matching--Miss Piggy and music are the reinforcements on this disc.

We use many kinds of sensory materials with Ro. All materials reach more than one modality. A list follows:

Playdough

Colored orange: scented with orange koolaid or flavoring
green: scented with mint flavoring
brown: scented with root beer flavoring
white: scented with coconut flavoring
yellow: scented with banana flavoring
yellow: scented with lemon koolaid or flavoring
red: scented with strawberry koolaid or flavoring
red: scented with cherry koolaid or flavoring
purple: scented with grape koolaid

Sensory input:

lotion
powder
shaving cream
whipped cream
wet and dry sand
bird seed
oats
rice tinted with food coloring
potting soil
instant potatoes
sunflower seeds
popcorn
lima beans
large buttons
cedar chips
snow (when available)
plastic straw
shells
baking soda
potter's clay
small pine cones
cotton balls
plastic bubbles for popping (found in packing materials)
confetti
egg shells
goop (cornstarch & water)
corn meal
fine salt
rock salt
sugar
colored gravel
foam
grits
dried peas
macaroni (all shapes)
barley
dried kidney beans
dried pinto beans
bread crumbs
cooked noodles (tinted)
cornstarch
cooked spaghetti (tinted)
nuts (all types in shells)
fabric (velvet, silk, wool, cotton)
grass seed
tub of water (tinted blue with sea shells on bottom)
water with dish soap for bubbles

Fingerpaint:

with vanilla, lemon or chocolate pudding

with fine salt, coarse salt or sand

Roselyn

Submitted by Klass Jagersma and Heather Bergsma
Dufferin Christian School
Carman, M.B.
Canada

Roselyn is a 12 year old girl. She began to regress at approximately 12-15 months of age. It wasn't until last year that she has been diagnosed as having Rett sybdrome. For four years Roselyn was in a Kindergarten program for 3 days a week with the other 2 days on a one on one with a teacher. The program was less and less meeting Roselyn's needs and became less and less age appropriate. At the moment Roselyn is in her second year of full integration into a grade 4,5 combination at a parental Christian elementary school.

Roselyn can walk along when supported with one hand or with the help of a walker. She stands independently for short periods of time (up to 2 minutes) and sometimes takes a few steps on her own (4-5). With eating and drinking she reaches for the cup, or food and often attempts to bring it to her mouth. She has trouble with her fine motor skills (pincer grip) and needs help to successfully bring it into her mouth.

She has been toilet trained during the day through habit-training. Roselyn is non-verbal, but does make several sounds indicating her moods: cry, laugh, whine, yell. At the moment we are trying to teach Roselyn to communicate through eye pointing. She reacts very well to familiar voices by looking up or smiling. Especially certain records (children's songs and country music) have a very stimulating influence. Also, Roselyn responds very well to a firm voice. She loves to have one hand in her hair while at the same time sucking the thumb of her other hand. Quite often she wears a mitt to fight this habit. Verbal prompt, snap finger and physical prompt are used to which Roselyn responds very well (however, left alone she continues those habits.)

Roselyn is in a 24 hour program in which all programs are carried out each day at home and at school. Combined with this is her enrollment in a fully integrated setting.

Two important aspects of Roselyn's program are the physio-therapy and communications. Twice daily (once at home and once at school) Roselyn has physiotherapy with full "range of motion" (each joint is stretched to full capa-city) and balance exercises. This is all reinforced by the students who involve her in the day to day activities in the school.

During communication Roselyn and the teacher's aide work on eye-pointing, imitation by means of the Echo-system and other forms of responding to the environment. This also is reinforced all day by the students and the family members. This tremendous active involvement all day is the key to Roselyn's social, mental and physical well-being.

Lindsay

Submitted by Diana Spencer
Cheryl Ann School for Mental Retardation
and Developmental Disabilities Program
Celina, Ohio

The following is a description of the pre-school program implemented for Lindsay, a five-year-old Rett syndrome girl from Celina, Ohio:
Lindsay is a 5 year old girl who attends the pre-school program at Cheryl Ann School - a county program for the Mentally Retarded and Developmentally Disabled, serving clients with infant stimulation, pre-school, school-aged, and sheltered work programs.

She is at a point in the progression of Rett syndrome where her regression has apparently slowed and we are seeing some progress in regaining lost skills. Except for the fact that she didn't walk, her development was rather unremarkable until around 18 months. At 18 months she began seeing specialists to determine the cause of her difficulties in ambulation. At about the time of her second birthday, more severe problems manifested themsleves. She had her first seizure; she began hours of screaming and crying without apparent cause; language and social interaction were lost. By the time she entered our pre-school program at three years, she had lost most purposeful use of her hands, although she still finger fed herself. Her screaming and tantrums were lessening but she seemed disinterested in people and objects. At present, she walks with an extremely broad-based, stiff-legged gait with one or both hands held (she never walked independently except for a few steps only days before her sudden regression began). She is totally dependent in feeding, dressing, and is not toilet trained. She is good natured, smiles appropriately and is interested in people and activities around her.

GROSS MOTOR SKILLS:

Lindsay has developed a slight scoliosis due to poor muscle tone in her back. Her orthopedist recommended a supportive chair so she now has a wheel chair to provide support and assist in travel. The chair, however, is not used to travel short distances such as from room to room within the school or at home.

At one point she began to resist changes in the texture under her feet (e.g. she stopped at the door of our room and would not walk from carpet to the tile floor). After a program of deep massage to her soles and a program which required her to walk barefoot on different textures, she now shows no reluctance to walk on various surfaces. She receives physical therapy twice monthly and her exercises are carried out daily. Most of her exercises involve weight shift, balance, and work on her tone. Many of these are done on a large therapy ball.

Lindsay has maintained extremely good eating skills. She chews well and can eat virtually any foods (she especially enjoys crunchy foods like Fritos). Last year we were able to teach her to drink through a straw which increased her fluid intake considerably. We use a cup with a lid to reduce spills and use aquarium air line tubing rather than traditional straws because it is sturdy and reusable and also is clear so it is easier to monitor whether or not she has gotten any fluid.

She has always relied on others to break up her food in small bites and place it in her mouth. This summer, however, we used basic jaw-control techniques to teach her to bite off pieces of cracker for herself. We are currently working on more difficult biting on sandwiches, etc.

Lindsay is not toilet trained, although we plan to evaluate her and attempt a clock-training program in the near future.

FINE MOTOR:

Lindsay currently cannot hold anything in either hand. We have attempted to get purposeful gross movement from her arms and hands, however. We have done some informal activities with large switches she can strike or push on in order to turn on various appliances such as stereos and lights and battery-operated toys.

We recently have been encouraged about a program we have begun which allows her to indicate wants and preferences at meal time by touching the food/drink she wants. We began by putting her cracker on one side of her and her glass of juice on the other. We then took her hand and touched the item with it before we gave her a bite or drink. We have only worked on this skill a short time this school year but she seems to understand what we expect when we now say "Do you want more cracker? Show me." She is making the touch on her own now with fairly high consistency. We hope that success in this area can lead to better communication skills in many areas of her program.

She also receives deep massage and tactile stimulation to her palms in order to maintain the good extension and range in her hands in addition to making her less defensive and more likely to use her hands purposefully.

SOCIALIZATION:

I feel it is important to mention in this outline of Lindsay's program the important role her interaction with other students has played. In her short time in our program, she has been in two very different types of classroom settings. Prior to this year, she was in a "Developmental Class" which served the most profoundly handicapped

children in the program in terms of both physical and mental disabilities. The class had one certified teacher and two full time assistants for eight students. The program was based on one-to-one experiences for the child and her teacher. In Baltimore I heard parents who were very outspoken in their demand for such a program for their daughters. Some went so far as to refuse to enroll their child in school until the school agreed to such a placement. This year our pre-school program is much more integrated with motor and cognitive skills which range from approximately the one to two month level to those who have only very minor delays and are very likely to be on their way to public school programs by the time they reach school age. It has been my experience that the latter has been much more beneficial to all students involved, including Lindsay and even those whose handicaps are more severe than hers.

In the first arrangement I mentioned, the students do get one-to-one attention. However, in a class of such severely handicapped children the interaction and language is virtually non-existent. In the integrated pre-school, students are constantly bombarded by language, touches, and affectionate interaction. I can say that even our most involved student never sits for longer than 10 minutes without an ambulatory child taking his hand, saying his name, putting a toy in his hand or giving him a kiss on the head! The animation in Lindsay's face when she is between two boys sending a wind up train back and forth was never seen in the more sterile setting of the segregated program of years past. I may add, also that with careful planning Lindsay and her classmates are receiving just as much individualized instruction as in the "Developmental" program. See, you can have the best of both worlds!

Angie

Submitted by Gay Naganuma, P.T.
University of Washington
Experimental Education Unit
Seattle, Washington

Angie is sixteen years old and attends the Experimental Education Unit in a classroom for students with severe and profound handicaps. In addition to two educators, the classroom is staffed by a physical, occupational and speech therapist. Angie has six classmates, including two other girls who have Rett syndrome.

Angie can ambulate independently and tolerates uneven surfaces. She had scoliosis surgery several months ago and her gait and pattern has improved dramatically. She can go up and down stairs independently, but requires mimimal assistance for her rate to be functional. All she needs is help with advancing her hand on the rail to get her to take the next step. Occasionally, a touch prompt behind her knee is needed to get Angie to flex that lower extremity. Angie can also sit down and come to stand from a chair independently.

Angie's hand usage includes finger-feeding bite-sized pieces of food, eating with a spoon with occasional help to scoop, picking up and drinking from a cup and pressing a switch to activate a toy or record player. She began wearing bilateral thumb abduction splints a few months ago and they have been successful in reducing the amount of time she spends engaged in stereotypic hand movements. The splints have not stopped the behavior, but have reduced it to a point where her hands are apart enough to allow implementation of programs working on functional hand use.

Facilitation of equilibrium reactions is emphasized in therapy both in sitting and standing positions. Sitting equilibrium responses are worked on using a large therapy bolster or ball. In standing, she tolerates several minutes on a tiltboard, shifting her center of gravity laterally and in an anterior/posterior plane. Practice of transitional movements is also emphasized (prone to sidelying to sidesit to quadruped to full kneel to half kneel to stand).

Changing positions sometimes seems threatening to Angie, so sessions go most smoothly if movements are done slowly, if she is told what is coming next and if we take frequent breaks for Angie to rest and calm herself. We also practice using a "steppage" gait pattern (simultaneous hip and knee flexion) to improve her stair climbing ability. Angie tends to shuffle her feet, so stepping over 6-9" hurdles encourages her to really shift her weight and take a step rather than a "slide". Therapy sessions also incorporate lots of upper extremity weight-bearing prone over a therapy ball or in sidesit position.

Angie is very social and would rather interact with a person than with a toy. She loves hugs, music, Sesame Street, going for walks and Teddy Ruxpin. It is very easy to get Angie to smile. Actually, it is pretty easy to get Angie to laugh out loud!

Submitted by Sheila Merzer and Lyle Chastain
Minneapolis Children's Medical Center
Minneapolis, Minnesota

In 1974 while consulting in a school district in Illinois, Lyle Chastain and Sheila Merzer saw a young girl sitting in a special education classroom wringing her hands together in a way that seemed very familiar. Sheila and Lyle asked the teacher about this child and were able, without looking at the records, to guess this child's early history which included normal development until age two.

This was the seventh child with a similar history and similar presenting characteristics that they had seen, and it was from this point on that Sheila and Lyle began referring to these young girls as the "Seven Little Girls Syndrome" at educational workshops and medical staff training. It wasn't, however, until 1983 when Dr. Reno Backus, a pediatric neurologist who works closely with Exceptional Children with Communication and Interaction Disorders, a program at Minneapolis Children's Medical Center, came across the article by Bengt Hagberg that the official diagnosis of Rett syndrome was given to these children.

In the late 1970's Sheila and Lyle were actively involved in programming for approximately 12 of these children throughout the midwest. At that point in time, within most of the children's classrooms, there was considerable emphasis on stopping the hand stimulation movements of these children. Lyle and Sheila actively advocated for these children and, in several cases, were able to prevent program staff from implementing such aversive procedures as squirting the child in the face with vinegar water solutions in order to stop the self stimulatory behavior. Instead, they put together a series of activities geared toward helping the children use their hands in a more positive and productive way and helping staff learn to work with these children without writing goals that were impossible to reach. In many cases, changing staff attitude toward the children was much more relevant than the actual program design provided for the children's classroom activity.

In addition to working with staff in developing an understanding of these children's patterns and functioning, Sheila and Lyle developed a variety of cause-effect activities geared toward maximizing the children's functioning. These activities generally consisted of cause-effect sequences designed around materials and interactions which these children found intrinsically reinforcing. As much as possible, gross motor and kinesthetic input was incorporated within the activity sequence. For example, a child might be encouraged to hold out her hand to receive powder or lotion, then touch the adult's hand to indicate she wanted the lotion rubbed on her hand. A child might also be helped to follow a simple obstacle course to find a desired item (e.g., piece of food) at the end. Use of hands is encouraged within these activities by enabling

a child to experience a successful environmental interaction simply by reaching out and touching, for example, a see-and-say to get the teacher to pull the string or holding on to the sides of a wagon in order to get the teacher to pull the wagon.

Other activities such as tying a child's favorite toy to the end of the string and helping the child pull the string in order to get their favorite toy, helping a child push a popper toy or a doll carriage across the floor, having a child touch a musical toy or instrument and having an adult facilitate that instrument making a sound, hiding objects such as raisins under cups for the child to pick up and find, etc. have all proved to be helpful in various cases.